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SPLENECTOMY IN AN ABNORMAL COAGULATION PROFILE PATIENT WITH EXTRA-HEPATIC PORTAL VENOUS OBSTRUCTION

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ABSTRACT

The clinico-surgical implication and successful management of a case of an abnormal coagulation profile patient withportal hypertension and splenomegaly due to extra-hepatic portal venous obstruction is presented.

A 14-year old girl had gastro-esophageal variceal bleeding and splenomegaly. During preoperative workup prolonged prothrombin time and severe thrombocytopenia were detected. Splenectomy with devascularisation was successfully performed with transfusion of fresh frozen plasma and platelet rich plasma in the perioperative period. At surgery there was no excessive bleeding. The perioperative course was uneventful and the patient is doing well on follow up.

Surgical portal decompressive procedures can be safely undertaken in an abnormal coagulation profile patientswith portal hypertension if meticulous surgical hemostasis is achieved at operation and the deficient blood products are adequately replaced in the perioperative period.

KEYWORDS: Extra hepatic portal venous obstruction, Splenectomy, abnormal, coagulation profile.

INTRODUCTION:

Extra hepatic portal venous obstruction (EHPVO) characteristically refers to obstruction of the trunk of the portal vein, its branches and may even extend to splanchnic veins. Among the etiology of EHPVO, it can be infection and/or inflammation like omphalitis, neonatal umbilical sepsis, intra-abdominal infection, post umbilical catheterization or it could also be portal vein injury, developmental anomaly, prothrombotic causes, and idiopathic. Pathogenesis of EHPVO includes presinusoidal blockage of the portal vein, normal intrahepatic venous pressure, raised intrasplenic pressure, hypersplenism and opening of portosystemic collaterals (Wani, 2015). Patients with EHPVO most commonly present with upper gastrointestinal bleeding splenomegaly and pancytopenia. (Vogel, 2017). In children, 70% of all variceal bleeds are due to EHPVO.Major concerns in such cases are hypersplenism, variceal bleed, growth retardation, decreased lean body mass and loss of muscle. (Sarin, 2006). Massive splenomegaly causes the destruction of the pooled blood cell components leading to thrombocytopenia, leucopenia, and anemia making the child susceptible to recurrent infections. Due to multiple transfusions in the past patients may develop atypical antibodies. (Govil, 2018).

MATERIALS AND METHODS:

A 14 year old girl was admitted in the department of GI and general surgery, Tribhuvan University Teaching Hospital (TUTH) with the diagnosis of Extra Hepatic Portal Venous Obstruction (EHPVO). She gave history of abdominal fullness with discomfort which was increasing since the age of 7 year, yellowish discoloration of eyes on and off, high grade fever with chills and rigor, frequent spells of dizziness and weakness. However there was no history of bleeding, petechae, bruises, nausea vomiting, and shortness of breath. Since last 2 months she was admitted at children's hospital for fever with chills and rigor, upper GI bleeding for which she gave history of transfusion with multiple units of blood and blood products and was referred to our hospital for further management. On clinical examination she was thinly built, pale, icteric, weighed only 26kg. Her vitals were stable except for tachycardia (pulse rate 110/min). The systemic examination was within normal limit except per abdominal examination revealed splenomegaly extending 6 cm below costal margin, it was non-tender and smooth. Airway assessment was normal. Blood investigations revealed pancytopenia due to splenomegaly: hemoglobin (Hb) 10.0 g/dL, total leucocyte count of 780/mm^{3}and platelet count 15,300/mm^3. Liver function tests showed prolonged prothrombin time (PT) - 20sec,INR – 1.8, serum bilirubin: total - 80.0 uMol/L, direct - 15.0uMol/L, serum alkaline phosphatase: 464 U/L, serum aspartate transaminase: 41 U/L, serum alanine transaminase: 25 U/L. Other routine investigations were within normal limit. UGI endoscopy revealed 2 columns of large and 1 column of small esophageal varices with portal hypertensive gastropathy and was advised for endoscopic variceal ligation if Splenectomy was not planned. Her contrast enhanced computed tomography (CECT)of abdomen showed features suggestive of extrahepatic portal vein obstruction with huge splenomegaly (26 cm), multiple collaterals including lieno-renal shunt.

The possible problems during surgery with regard to excessive bleeding were explained to the parents and an informed written consent was obtained. A clinical haematologist saw the patient. She was given iv vitamin K and transfused 350ml of FFP before the start of the operation and repeat PT showed 18sec with INR 1.6. Other blood and blood products were kept ready. Essential monitoring HR, ECG, SPO2, non-invasive blood pressure monitoring was done. Two intravenous access with 20G iv cannula was obtainedand induction of anesthesia with propofol, fentanyl, and cisatracurium was done. Laryngoscopy and intubation done with 6 mm ID endotracheal tube. Anesthesia was maintained with oxygen and isoflurane. One unit of Platelet rich plasma (PRP) was given during induction and other two units during surgery. Splenectomy and devascularisation was done. The operative time was 4 hours and the patient remained hemodynamically stable throughout the procedure. Patient also received 1 unit whole blood after removal of the huge spleen though intraoperative blood loss was only 200ml. At the end of a surgery, the patient was extubated after complete reversal of anesthesia effects and muscle paralysis. Patient shifted to post operative ward for monitoring for 24 hours before moving to the ward. She received 600ml of FFP in the postoperative ward as her PT increased to 29sec and INR 2.6.There was no bleeding from the drain sites and the wounds healed well without haematoma formation.

Patient's postoperative stay was uneventful and she was then discharged on the eighth post- operative day with investigation reports which suggested no evidence of hypersplenism (hemoglobin- 12.9gm/dl, total leukocyte count-5700/mm^3, platelets- 163,000/mm^3) and PT/INR – 17/1.5.

DISCUSSION:

Extra hepatic portal venous obstruction (EHPVO) is common in the developing countries. 70% of pediatric patients with portal hypertension have EHPVO. But in West it is the second cause after cirrhosis. EHPVO is a common cause of upper gastrointestinal bleeding in children. (Sarin, 2006). In congestive splenomegaly, the spleen volume can be 10 times greater than normal, with 30-90% of platelets and 40% of erythrocytes located in the spleen.

Anaesthetic concern in management of our case was severe low platelet count and increase prothrombin time (PT). Increase in PT was corrected by preoperative transfusion of FFP. But dilemma was whether to correct low platelet count preoperatively or not. Was elevation of platelet count needed by transfusion before procedure? If needed, then how much of platelet count is safe for surgery and anesthesia?

Platelet count at which any procedure can be safely done is unknown. Benefit and risk of perioperative platelet transfusion should be assessed on a patient by patient basis. A normal platelet count is 15-40 times higher than is necessary to achieve hemeostasis. (Slichter, 2004). Platelets are involved not only in haemostasis, but also in inflammatory and immune responses and wound healing. (Li, 2011). In pathologic condition, platelets may promote excess inflammation and be associated with organ damage, like acute kidney injury and acute lung injury. (deStoppelaar, 2014).

Thrombocytopenia has been subcategorised for surgical purposes into mild (100000-149000/mm^3), moderate(50000-99000/mm^3), and severe (< 50000/mm^3). (Williamson, 2013). Besides platelet count, many other factors like

increasing age, (Cortelazzo, 1991), aetiology of thrombocytopenia, perioperative administration of fluids, blood products, medications used affect the haemostatic effectiveness of a given platelet count.

No specific guideline is present about platelet count for airway management. Use of video laryngoscopy allows easy and successful intubation with less traumatic intervention. As we did not have the facility of video laryngoscope, we used Macintosh laryngoscope to intubate without causing any trauma to the airway. No specific interaction is known with any anesthetic agents. Use of NSAID should not be used in patient with thrombocytopenia as it affects platelet function. Veen et al (Veen, 2010) reviewed the current guidelines, case series and case reports on epidural and spinal anesthesia and lumbar puncture, this review reported that platelet count of 80000/mm^3 is safe for placing or removing an epidural or spinal anaesthetic and 40000/mm^3 is safe for lumbar puncture. Consistently low platelet count is less problematic than rapidly falling values due to other diseases, which are often accompanied by platelet dysfunction or coagulopathy. Neuraxial anaesthesia was successfully performed in several studies with platelet counts between 50000 and 80000/mm^3. (Spahr, 2008) (Govil, 2018). A study done in Spain with 199 patient showed laparoscopic Splenectomy (LS) done at platelet counts between 10000 and 50000/mm^3 was no different from LS at counts greater than 50000/mm^3, in terms of blood loss, operative time or postoperative complications. However, platelet counts lower than 10000/mm^3 had an important impact on perioperative outcome, with significantly greater intraoperative blood loss, longer operative time and prolonged hospital stay. (Bele, 2016).

Gazula et al (Gazula, 2009) did a study in 28 patients where they have shown that early ligation of the splenic artery during Splenectomy results in passive splenic decongestion and thereby a significant gain in blood components. The gain in hemoglobin and hematocrit was equivalent to a transfusion of at least 100-150 ml of packed RBC, platelet transfusion of at least 4 units of platelet concentrates in an adult. This protocol appears to be a feasible blood conservation method to avoid blood transfusions in this group of hypersplenic EHPVO patients.

CONCLUSION:

During Splenectomy in EHPVO patient, even with low platelet count (10000-50000/mm^3), blood loss need not be a concern as the platelet count at which a given procedure carries an acceptable risk of bleeding is unique in each patient. The true magnitude of benefit of prophylactic platelet transfusion remains unclear and the need for platelet transfusion should be assessed on a patient-by-patient basis. Need of blood product transfusion can be minimized with meticulous surgery, early ligation of splenic artery and passive splenic decongestion.

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